

Bilevel positive airway pressure therapy in a patient with myotonic dystrophy and postoperative respiratory failure: A case report

ABSTRACT

Respiratory failure is a common complication in patients with myotonic dystrophy (MD) and might be a presenting symptom in the perioperative setting. We report the case of a 59-year-old woman with MD who underwent open cholecystectomy and developed postoperative respiratory failure. Without reintubation, the patient was successfully managed with bilevel positive airway pressure (BiPAP) and was discharged uneventfully. BiPAP may be considered as an alternative for postoperative respiratory failure in patients with MD. Careful observation of patients' postoperative condition and an earlier application of BiPAP are instrumental in avoiding retracheal intubation, which may cause further serious problems in patients with MD.

Key words: Bilevel positive airway pressure; myotonic dystrophy; postoperative respiratory failure

Introduction

Myotonic dystrophy (MD) is a chronic and slowly progressive disease that can manifest at any age from birth to old age.^[1] MD is one of the most common myotonic syndromes and is an autosomal dominant and a multisystem disorder involving skeletal, smooth, and cardiac muscle, posing substantial serious anesthetic problems.^[2] Because patients with MD have greater sensitivity to general anesthetics and muscle relaxants, serious perioperative problems have been noted, such as delayed recovery from anesthesia and perioperative cardiovascular and respiratory complications.^[2]

Respiratory failure is a common complication in patients with MD and may be a presenting symptom in the setting


of anesthesia or surgery.^[3] In patients with MD, respiratory failure, frequently caused by respiratory infection, is the major cause of death.^[3] Thus, respiratory management of patients with MD suffering from respiratory failure is challenging. To the best of our knowledge, there was no report regarding the effectiveness of bilevel positive airway pressure (BiPAP) therapy on postoperative respiratory failure, triggered by anesthesia and surgical stress, in a patient with MD. We here present the successful respiratory management of a patient with MD who developed late-onset postoperative respiratory failure using BiPAP.

Written informed consent was obtained from the patient for publication.

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Case Report

A 59-year-old female patient, body weight 60 kg and height 152 cm, was scheduled for laparoscopic cholecystectomy. A preanesthetic interview revealed that she had MD that was diagnosed when she underwent a Cesarean section 30 years ago. Her symptoms of MD included severe eyelid drooping, slight anarthria, and mild muscle weakness in the bilateral lower limbs [manual muscle test (MMT) 4/5], with neither grip myotonia nor dysphagia. She was able to continue walking for a distance of 100 m without rest. She was also diagnosed with type 2 diabetic mellitus, hyperlipidemia, and hypertension and is being treated with candesartan, atorvastatin, and metformin. Her preoperative laboratory test values were almost within the normal range, except for HbA1c of 9.0% and a blood glucose level of 245 mg/dL. Her chest radiograph and electrocardiogram showed no abnormalities. Her preoperative pulmonary function test showed a percent vital capacity (% VC) of 73.4% and a forced expiratory volume in one second (FEV1) of 85.4%. Her preoperative arterial blood gas analysis (BGA) at room air showed arterial oxygen tension (PaO₂) of 71.6 mmHg, carbon dioxide tension (PaCO₂) of 50.9 mmHg, and pH of 7.432.

No premedication was given. Thoracic epidural catheter insertion in the 9th and 10th thoracic interspace was performed in the right lateral decubitus position. General anesthesia was induced by propofol infusion with a target blood concentration of 3.0 µg/mL using a target-controlled infusion (TCI) device (TCI pump TE-371; Terumo, Tokyo, Japan). After loss of consciousness, fentanyl 50 µg was administered intravenously, and neuromuscular monitoring of the left ulnar nerve was commenced using the train-of-four (TOF) stimulus. Two minutes after administration of rocuronium 20 mg, all four twitch responses disappeared, and tracheal intubation was carried out with a 7.0-mm tube. During surgery, anesthesia was maintained with TCI of propofol at 2.0–2.5 µg/mL, supplemented by continuous remifentanyl infusion (0.05–0.1 µg/kg/min) and a continuous epidural infusion of 0.125% levobupivacaine at 4 mL/h. No additional rocuronium and fentanyl were given after anesthesia induction because stable muscle relaxation was maintained during surgery. Bispectral index (BIS) values were maintained between 40 and 50 during surgery. Because of severe inflammation and adhesion, the surgeons performed an open surgery instead of a laparoscopic cholecystectomy. At the end of surgery, the TOF showed four twitch responses, and the patient's spontaneous breathing showed a tidal volume of approximately 400 mL and a respiratory rate of 12/min. The neuromuscular block was reversed by administering 200 mg of sugammadex, and then we confirmed that TOF ratio was

more than 0.9. Her trachea was extubated uneventfully, and anesthesia time was 5 h 29 min. Her respiratory status on the ward was stable with 97% SpO₂ under O₂ administered at 6 L/min through a face mask.

On the postoperative day (POD) 1, despite the presence of a large amount of sputum, her SpO₂ was maintained at 96% in a sitting position under O₂ administered at 3 L/min through face mask. However, her SpO₂ decreased to below 88% without O₂ administration. Her arterial BGA under O₂ administered at 3 L/min showed PaO₂ of 68.9 mmHg, PaCO₂ of 58.2 mmHg, and pH of 7.349. On POD 3, due to further increase in sputum production, she complained of difficult sputum expectoration and dyspnea with tachypnea (over 20/min) and 85% SpO₂ under O₂ administered at 3 L/min. Her arterial BGA under 3 L/min O₂ showed PaO₂ of 53.1 mmHg, PaCO₂ of 67.3 mmHg, and pH of 7.364. Her consciousness level worsened further due to CO₂ retention. However, it was estimated that her respiratory failure and CO₂ retention had not reached yet the critical status. To avoid retracheal intubation, BiPAP therapy was applied using NIP Nasal V[®] (Teijin Inc., Tokyo, Japan) to support the diminished respiratory function. Initial settings included an inspiratory positive airway pressure (IPAP) of 10 cmH₂O and an expiratory positive airway pressure (EPAP) of 4 cmH₂O using a CPAP mask; O₂ flow of 2 L/min was delivered via the mask. On POD 4, her arterial BGA under BiPAP showed PaO₂ of 77.9 mmHg, PaCO₂ of 54.6 mmHg, and pH of 7.436. Her respiratory status significantly improved. BiPAP therapy was continued from POD 3 to POD 5. Her SpO₂ maintained 96%–97% under BiPAP therapy. On POD 6, she was weaned off BiPAP because of no dyspnea and significant decrease in sputum. Under O₂ administered at 0.5 L/min via nasal cannula, she breathed smoothly at a respiratory frequency of 12–14/min without any complaints, and her SpO₂ was maintained at 96%. On POD 7, she started oral food intake and walking in the ward. On POD 9, she achieved a 50-m walk without rest in the ward. On POD 10, she was discharged from the hospital. Her last vital signs included blood pressure 98/55 mmHg, heart rate 72/min, body temperature 36.8°C, and SpO₂ 95% at room air.

Discussion

Respiratory failure often occurs with the progression of the muscular manifestations of MD during the course of the disease.^[3] There were some reports describing the perioperative management of a patient with MD, who developed postoperative respiratory failure.^[1,4-6] Most patients with MD and postoperative respiratory failure were reintubated and mechanically ventilated until their respiratory conditions significantly improved and met the

extubation criteria.^[1,4,5] Noguchi *et al.* reported successful respiratory management in a patient with MD using high flow nasal cannula therapy, which has been recently introduced as an innovative respiratory support for critically ill patients.^[6]

Unfortunately, there is still no randomized trial to compare noninvasive ventilation (NIV) with invasive ventilation in acute neuromuscular respiratory failure.^[7] Thus, the effectiveness of NIV methods such as BiPAP in these conditions is controversial. However, Shneerson *et al.* suggested that NIV may be effective for respiratory failure in a patient with neuromuscular disorder because it is a well-established therapy for these conditions.^[8] Furthermore, Chau *et al.* reported that infants with congenital MD suffering from respiratory failure were successfully managed with BiPAP.^[9] In addition, Souayab *et al.* reported that adult patients with previously undiagnosed MD, who developed acute respiratory failure not triggered by anesthesia or sedation, were successfully managed with BiPAP.^[3] Accordingly, NIV such as BiPAP may be effective in postoperative respiratory failure with MD. Basically, NIV improves oxygenation and reduces respiratory muscle fatigue in critically ill patients.^[10,11] We inferred that in the present case, respiratory collapse was caused mainly by difficult sputum expectoration due to respiratory muscle fatigue, triggered by surgical stress and anesthesia. Therefore, we considered BiPAP therapy, which reduces respiratory muscle fatigue, resulting in easy sputum expectoration, promoting CO₂ washout and better oxygenation. Therefore, BiPAP therapy was chosen to avoid retracheal intubation in this case. The most important reason for avoidance of tracheal intubation was that patients with MD have greater sensitivity to general anesthetics,^[2] which is necessary for sedation in intubated patients under mechanical ventilation. Thus, respiratory management under tracheal intubation in these patients may cause further serious complications, such as delayed recovery from sedative medications, ventilator-associated pneumonia, and cardiovascular complications. Therefore, careful observation of patients' postoperative condition and an earlier application of BiPAP therapy in such cases may be one of the most important factors in avoiding retracheal intubation, which may cause further serious problems perioperatively. However, the patient's optimal cooperation and tolerance are required for BiPAP therapy because the CPAP mask must be tightly applied on the patient's face. Therefore, BiPAP may not be appropriate

in cases with poor compliance, dementia, and mental retardation.

In conclusion, an earlier application of BiPAP therapy can be recommended in patients having MD who develop postoperative respiratory failure. Further clinical studies are warranted to test the efficacy of NIV in an acute neuromuscular respiratory failure.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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